Newborn Screening Follow-up for Isolated C5 Acylcarnitine Elevations (also applies to any plasma or serum C5 acylcarnitine elevations)*

Did patient receive antibiotic treatment (ie, ampicillin)?

**NO**

- Plasma or serum C5–High
- Urine isovalerylglycine–High
- Urine 3-methylbutyrylglycine–Normal

**YES**

- Likely pivalic acid (antibiotic) artifact

Perform:
- FAO / Fatty Acid Oxidation Probe Assay, Fibroblast Culture
- FIBR / Fibroblast Culture

Diagnostic for isovaleryl-CoA dehydrogenase deficiency

Did patient receive antibiotic treatment (ie, ampicillin)?

**NO**

- Isovalerylcarnitine–High
- 3-Methylbutyrylglycine–Normal

**STOP**

Call for consultation to determine the necessity for molecular testing:
- IVDA / Isovaleryl-CoA Dehydrogenase (IVD) Mutation Analysis (A282V)
- Isovaleryl-CoA Dehydrogenase (IVD) Sequencing

**YES**

- Diagnostic for short/branched chain acyl CoA dehydrogenase (SBCAD) deficiency
- Consider confirmatory testing for SBCAD gene

Perform:
- OAU / Organic Acids Screen, Urine
- ACYLG / Acylglycines, Quantitative, Urine
- ACRN / Acylcarnitines, Quantitative, Plasma
- ACRNS / Acylcarnitines, Quantitative, Serum

Routine labs performed locally: Glucose, electrolytes, blood gas, ammonia

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- Plasma or serum C5–High
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- Urine 3-methylbutyrylglycine–High

* Interpretive report provided for all tests in this algorithm